CYSTIC FIBROSIS

ORGANIZATIONAL GUIDELINES

CYSTIC FIBROSIS[1] Organizational Guidelines represent the minimum requirements for providing care for individuals with Cystic Fibrosis. Care and treatment should be provided in a manner, which includes adherence to and consistency with each of the following Guidelines.

CRS Enrollment:

All members with a cystic fibrosis diagnosis must be enrolled in clinics meeting these Guidelines. Care provided to members under CRS in any other part of the state must be coordinated with the designated Cystic Fibrosis Team.

Interdisciplinary Team Membership:

The following Team Members must attend clinics and team conferences, review patient information, determine the need to see the patients at a clinic site and are available for inpatient consultation or coordinate care with inpatient staff. It may not be necessary for each member to see the patient at each visit:

- Pulmonologist ï Lead Physician
- Registered Nurse Coordinator
- Social Worker
- Respiratory Therapist
- Dietitian/Nutritionist
- Child Psychologist
- CRS Member / Caregiver
- Primary Care Physician[2]

Available Personnel:

Below are personnel who must be available to the member / adult at the clinic during a scheduled specialty clinic. These personnel may or may not be called in by the Interdisciplinary Team members to see the member or they may choose to see the member based on prior knowledge of the members needs:

- Advocate

[1] These Guidelines of care are consistent with the Cystic Fibrosis Foundation, Minimum Criteria for CF Center Qualifications with minor revisions to reflect the Childrens Rehabilitative Services delivery system.
[2] The Primary Care Physician will be invited to all Team meetings; however, it is understood that PCP will not always be able to attend.

- Child Life Specialist
- Educator
- Translator

Consultative Personnel:

The clinic must have access to personnel for consultation including but not limited to the following:

- Allergist / Immunologist
- Anesthesiologist
- Angiographer
- Cardiologist, pediatric and adult
- Endocrinologist
- Gastroenterologist
- Geneticist / Genetic Counselor
- Infectious Disease Specialist
- Internist
- Nuclear Medicine specialists
- Otorhinolaryngologist
- Pediatrician
- Pulmonologist
- Radiologist
- Surgeon, pediatric, general thoracic
- Urologist

Outreach Clinics:

Outreach Clinics are designed to provide a limited specific set of services including evaluation, monitoring and treatment in settings closer to the family than a clinic. Major treatment plan changes must be communicated to the clinic. Cystic Fibrosis Outreach / Field Clinics must include the following personnel:

- Pulmonologist
- Clinic Coordinator
- Respiratory Therapist

Community-based Services:

Community-based services means all local services including provider agencies, schools, private physician offices, hospitals, and/or any other local setting. The following community-based services may be provided for patients with cystic fibrosis:

- Pharmacy services
- Respiratory therapy
- Lab work
- Any other appropriate extension of services as approved by the Interdisciplinary Team and approved by the CRS Medical Director.

Services to Adults with Cystic Fibrosis:

1. Clinic times separate from the member clinics will be scheduled for adults.
2. Child Life Specialist should be removed from the Team.

Facilities & Services:

1. Age-appropriate setting for adult patients
2. Defined age-appropriate services/ i.e. Pediatrics, Adolescent Medicine and/or Internal Medicine
3. House Officers when the hospital is a teaching institution
4. Pediatric and Adult Intensive Care Units
5. Respiratory Care Department
6. Nutrition or Dietary Department
7. Social Work Department
8. Identified clinic area
9. Laboratories performing:
    a. Sweat test by quantitative pilocarpine iontophoresis as stated in the Cystic Fibrosis Foundation guidelines
    b. Pulmonary function tests, including ability to measure long volumes
    c. Daily, round the clock availability of:
        i. bacteriology
        ii. blood chemistries
        iii. arterial blood gases
        iv. imaging studies including radiographic, ultrasonographic, and nuclear medicine studies.
10. Respiratory therapy available 24 hours a day, 7 days a week

Other Criteria:

1. Written patient records to include, but not limited to:
    a. Documentation of positive sweat test and/or genetics
    b. Copies of outpatient clinic visits
    c. Discharge summaries
    d. Growth chart
    e. Laboratory data
2. Sweat Test results reviewed by Lead Physician
3. Referral plan for procedures or services not available through CRS, such as oxygen at home, equipment, supplements, therapy vest, etc
4. All outpatient clinic reports and discharge summaries sent to the referring physician
5. A plan for the care of adult patients
6. 24-hour coverage by a lead physician
7. A minimum of 50 CF patients

Team/Staff Meetings:

1. Case Planning Meetings- a meeting of the specialists involved in the care and treatment of the member is to be held after each specialty clinic
2. Interdisciplinary Team Meetings/ review and planning meetings (patient specific):
    a. Every three (3) months with the regular assessment
    b. Once a year for planning and review with the family
3. Staff meetings at least annually to focus on issues of clinic patient care and clinic administration
4. Internal education meetings annually to focus on new information regarding the care and treatment of persons with Cystic Fibrosis
5. A yearly note by Social Services, Dieticians, and Child Life required

Lead Physician Specialists:

Qualifications: The lead physician specialists for members with cystic fibrosis will be a pulmonologist or pediatrician with experience in the care of cystic fibrosis patients. Board certification in Pulmonary Medicine is recommended.

GUIDELINES FOR PATIENT SERVICES, EVALUATION AND MONITORING FOR CYSTIC FIBROSIS

The purpose of these guidelines is to promote a uniform level of care and teaching services at CF Centers, and to provide a general framework for good patient care. Their relevance to specific situations will depend on individual variations in clinical course and

professional judgment. In addition, this document should serve as a tool to assess programs, secure resources needed to enhance patient care and education, and guide the future growth and development of CF care.

Diagnosis:

Goal: To provide accurate and timely diagnosis of CF.

The diagnosis of cystic fibrosis is based on clinical and laboratory findings. These may include but are not limited to: a) chronic obstructive pulmonary disease, b) intestinal malabsorption, c) electrolyte loss through sweat, d) family history of CF, e) meconium ileus at birth, f) male infertility due to azoospermia, g) presence of staphylococcus aureus or mucoid pseudomonas aeruginosa in the respiratory tract.

Confirmation of the diagnosis currently requires two positive sweat tests, done on different days, by quantitative pilocarpine iontophoresis (Gibson-Cooke) according to Cystic Fibrosis Foundation approved methods or 2 CF mutations. A positive test is defined by sweat chloride measurements in excess of 60 milliequivalents per litre in an adequate sample of sweat (minimum of 75 milligrams in gauze or filter paper, or 15 microliters for the Wescor Collection system, collected over a 30 minute period). Repeat borderline sweat electrolyte measurements (40-60 meq/ml) require clinical correlation and judgment for diagnosis.[1]

Evaluation and Education of Newly Diagnosed Patients:

Goal: a) To provide accurate assessment of physical and emotional status, and to begin patient family education.

b) To help families cope with the emotional impact of diagnosis and formulate an appropriate therapeutic plan.

Evaluation of the newly diagnosed patient should include medical, nursing, nutritional, psychosocial, respiratory, and physical therapy assessments as well as laboratory evaluation and genetic counseling. A comprehensive education program must be developed to promote optimum understanding of the disease, adherence to treatment plans and adequate coping with the demands of chronic illness.

Ongoing Patient Evaluation and Monitoring:

Goal: To anticipate and treat physical and psychosocial problems and complications of the disease.

At least four visits per year to the Clinic are recommended. The number of visits will vary with factors such as age, degree of illness; time elapsed from diagnosis, and distance from a clinic. Use of clinical scoring at every visit is encouraged. In addition, interim visits to the primary care physician for general pediatric care are necessary. The primary care practitioner has an important role in administering immunizations, evaluation and treatment of milder pulmonary exacerbation in consultation with the CF Center physician, advocacy and assessment of family dynamics. Every patient should

[3]The CF gene and some of its mutations have been recently characterized. This finding is foreseen to contribute to confirmation of the diagnosis of CF in the future.

be encouraged to be seen on a regular basis at a CF Clinic and by a physician in the community.

Respiratory Evaluation and Therapy:

Goal: To achieve optimum respiratory status. To anticipate and treat progression and complications of pulmonary disease.

Complete respiratory history and examination should be obtained at every visit (including nasal examination for nasal polyps).

Spirometry is recommended to quarterly and more often if clinically indicated and during hospitalizations for pulmonary exacerbation. Spirometry is done at every visit at many centers. Complete pulmonary function testing (including lung volumes) should be done at least once a year (performed according to American Thoracic Society Guidelines) .[2]

Arterial blood gases or pulse oximetry needs to be done at least annually on patients whose forced expiratory value in one second (FEV1) is less than 40% of predicted normal and additionally, when clinically indicated (i.e. exacerbation, oxygen therapy).

Respiratory tract culture and sensitivity should be done at least four times per year but preferably at each quarterly visit, before initiation of intravenous antibiotic therapy and at sick visits when clinically indicated.

Chest roentgenogram should be obtained annually and on pulmonary-related hospital admissions. Scoring of radiographs is encouraged.

All clinics should have written protocols for managing respiratory complications (hemoptysis and pneumothorax).

Knowledge and performance of respiratory and physical therapy techniques should be evaluated annually.

Gastrointestinal System/Nutrition:

Goal: To anticipate and treat nutritional deficits and complications. The ultimate goal is to achieve optimum growth and nutrition.

Measurements should include height and weight, plotted on Guideline growth chart, for all patients, every visit. Other measurements such as triceps skin fold thickness and mid-arm circumference can be useful.

Nutritional assessment should be carried out annually and when there is evidence of weight loss or poor weight gain. This assessment should include, but not be limited to, protein, fat, carbohydrate, vitamin and mineral intake. Other measures include: a) Assessment of pancreatic enzyme and vitamin supplementation and measurement of albumin and/or prealbumin levels at diagnosis, and when indicated; b) Abdominal examination with particular attention to liver and spleen size and consistency; c) A protocol for the management of diabetes mellitus; d) Laboratory measurements to include evaluation of metabolic and liver status, complete blood count, and fat soluble vitamin levels.

[2] Am. Rev. Resp. Dis. 1987; 136; 1285-96

Psychosocial Issues:

Goal: To anticipate and treat social and emotional problems of patients and their families.

Psychosocial assessment should be carried out annually. The CF Clinic staff should be available for genetic counseling, crisis management, ongoing support and anticipatory guidance, when indicated. Sexuality, fertility and pregnancy should be discussed at age-appropriate intervals.

Adult Issues:

Goal: To ensure that the changing needs of the growing population of adult patients are met by caregivers.

Adult patients have specific needs different from those of the pediatric patients. Clinics should have clear plans for the care of adults including identification of appropriate caregivers and preparation for transition to adulthood and adult care. Adequate actions to address these needs include the incorporation of adult care specialists in the clinic program, establishment of inpatient services in internal medicine wards, creation of transition teams or creation of parallel adult care teams. The specific activities undertaken will depend on clinic size, geographic factors, local institutional idiosyncrasies and availability of specialists in the community.

Specific areas requiring services include: obstetrics and gynecology, urology, cardiology, endocrinology, adult nutrition, vocational counseling, independent living, family planning, sexuality, medical insurance and other special counseling.

Facilities:

Goal: To provide adequate inpatient, outpatient and laboratory facilities to meet special needs of the CF patient population.

These include a) inpatient facilities that provide separate quarters for older patients (i.e. adolescent ward or wing). Personnel trained in the care of CF patients and sensitive to their needs; b) In-house physician 24 hours per day, 7 days per week; c) A specified, identifiable outpatient area with adequate space (providing privacy) for interviews and consultations, team conferences, equipment for nutritional assessment, height/weight measurement, and other pertinent outpatient activities; d) Laboratories capable of completing pulmonary functions by ATS Guidelines, including body plethysmography, microbiology studies according to the Centers for Disease Control guidelines, sweat testing according to CFF guidelines, and blood gas determinations and non-invasive oxygen monitoring.